Unilateral absence of C1

Unilateral absence of C1 ranges from hypoplasia of a lateral mass to complete agenesis of a hemiatlas with rotatory instability and basilar impression. Two-thirds of the children present with symptoms at birth; the others develop torticollis and are noticed later. Neck flexibility is variable and decreases with age. The condition is not painful. Neurological signs are present in about a quarter of the patients. The natural history is unknown.

Standard anteroposterior and lateral radiographs rarely give the diagnosis, although the open-mouth odontoid view may suggest it. Tomograms or CT scans usually are needed to visualize the anomaly. Occasionally, there is an assimilation of the atlas.

The deformity should be observed to document the presence or absence of progression. This observation is primarily clinical (e.g., photographic) because radiographic measurements are difficult if not impossible to obtain. Bracing does not halt the progression of the deformity. Surgical intervention is recommended in those patients with severe deformities. A posterior fusion from the occiput-C2 or occiput-C3 with or without decompression is performed. The ideal age for posterior fusion is between 5 and 8 years, corresponding to the age at which the canal size reaches adult proportions [1)].

1)
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2656787/#CR32